TREATMENT RESPONSE OF HYDRONEPHROSIS ASSOCIATED WITH IDIOPATHIC RETROPERITONEAL FIBROSIS, FOCUSING ON RATIO OF IgG4/IgG3 SERUM CONCENTRATION

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Background: Hydronephrosis, a common complication of idiopathic retroperitoneal fibrosis (iRPF), may lead to poor renal outcomes unless it is resolved. Pathological confirmation can help to identify the aetiology of the disease and determine the treatment strategy. But, in most cases, it is difficult to obtain sufficient tissue due to the location of fibrosis. In a recent study, parts of iRPF are correlated with high-dose glucocorticoid treatment and impaired renal function.

Objectives: We aimed to investigate the ratio of IgG subclasses as a predictive factor associated with treatment response of hydronephrosis in patients with iRPF.

Methods: We retrospectively recruited 19 iRPF patients with hydronephrosis who evaluated serum IgG subclasses in a tertiary hospital between 2004 and 2019. Hydronephrosis was evaluated on the basis of imaging findings. Medications and clinical and laboratory findings, including IgG subclasses, were reviewed following the diagnosis of hydronephrosis. Hydronephrosis improvement on subsequent images was evaluated to assess treatment response. Categorised data were compared using chi-square or Fisher’s exact test. Continuous variables were compared using Mann–Whitney U test.

Results: At baseline, median serum IgG3 and IgG4 levels were 64 (IQR 37–82) mg/dL and 71 (IQR 40–171) mg/dL. Five patients had serum IgG4 levels >135 mg/dL and 11 patients had the reverse serum level of IgG4/IgG3. On subsequent images (median follow-up at 3.2 [IQR 1.7–4.0] months), 10 patients showed hydronephrosis improvement. The proportions of positive ratio of serum IgG4/IgG3 (81.8% vs. 25%, p = 0.024), periaortic involvement (81.8% vs. 25%, p = 0.024) and high-dose glucocorticoid treatment (45.5% vs. 0%, p = 0.045) were significantly higher in patients with improvement than in those without improvement (Table 1). Interestingly, even in cases with normal serum IgG4 levels, patients with improvement showed a higher serum IgG4/IgG3 ratio than in those without improvement (median 1.5 vs. 0.7, p = 0.038).

Table 1. Clinical characteristics and treatment according to the short-term outcome of hydronephrosis

<table>
<thead>
<tr>
<th>Improvement</th>
<th>No improvement</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Time to subsequent imaging (months)</td>
<td>2.2 (1.3–4.2)</td>
<td>3.2 (2.0–3.8)</td>
</tr>
<tr>
<td>Males (n, %)</td>
<td>8 (72.7%)</td>
<td>7 (67.7%)</td>
</tr>
<tr>
<td>Periaortic involvement (n, %)</td>
<td>9 (81.8%)</td>
<td>2 (25%)</td>
</tr>
<tr>
<td>Impaired renal function (n, %)</td>
<td>5 (45.5%)</td>
<td>2 (25%)</td>
</tr>
<tr>
<td>Serum IgG4 (mg/dL)</td>
<td>114 (59–172)</td>
<td>43 (36–109)</td>
</tr>
<tr>
<td>Elevated serum IgG4 (n, %)</td>
<td>4 (36.4%)</td>
<td>1 (12.5%)</td>
</tr>
<tr>
<td>Serum IgG4/IgG3 ratio</td>
<td>2.1 (1.2–4.9)</td>
<td>0.8 (1.0–4.0)</td>
</tr>
<tr>
<td>Positive ratio of serum IgG4/IgG3 (n, %)</td>
<td>9 (81.8%)</td>
<td>2 (25%)</td>
</tr>
<tr>
<td>Medical treatment (n, %)</td>
<td>7 (63.6%)</td>
<td>1 (12.5%)</td>
</tr>
<tr>
<td>High-dose glucocorticoid treatment (n, %)</td>
<td>5 (45.5%)</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>Surgical intervention (n, %)</td>
<td>8 (72.7%)</td>
<td>4 (54.5%)</td>
</tr>
</tbody>
</table>

a Values are median and interquartile range (25th–75th percentile)
b Defined as serum creatinine level > 1.24 mg/dL

Conclusion: The reverse ratio of serum IgG4/IgG3 was associated with hydronephrosis treatment response, thus suggesting favourable responses to high-dose corticosteroid.

References:

Disclosure of Interests: None declared

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THERAPEUTIC PROFILE OF PATIENTS WITH VOGT KOYANAGI-HARADA DISEASE FOLLOWED AT THE HOSPITAL DE CLINICAS, SAN LORENZO PARAGUAY

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Background: Vogt-Koyanagi-Harada Disease (VKHD) is an autoimmune eye disease that causes chronic vision complications in young people. The treatment is based on corticosteroids and immunosuppressants, to which biological drugs are currently added. The visual prognosis depends on the stage in which it is diagnosed and the effective and timely treatment.

Objectives: Describe the evolution and the therapeutic profile of patients with VKHD who go to the Rheumatology Department of the Hospital de Clinicas, San Lorenzo Paraguay until November 2019.

Methods: Descriptive cross-sectional study conducted at the Hospital de Clinicas, San Lorenzo Paraguay, where clinical histories of patients with this disease and who have regular follow-up were reviewed.

Results: 21 patients were included, 17 (81%) women, average age of 35 years. The diagnosis and treatment were made 15 (71%) in acute phase, 4 (19%) in convalescent phase and 2 (10%) in chronic phase.

Corticosteroids: All received corticosteroids at diagnosis. Methylprednisolone IV 1 g/day for 3 consecutive days in 8 (44%) cases and for 5 days in 10 (55.5%) cases. The first month everyone received prednisone orally at 1 mg / kg / d on average (66–75 mg/d) at 3 months average dose of prednisone was 37 mg / d, at 6 months 23 mg / d and at 12 months 10 mg / d / months.

Immunosuppressive treatment: They received 20 (95%) patients. Induction with monthly intravenous cyclophosphamide was performed in 8 (40%) cases and with Azathioprine in 12 (60%) cases. Of the patients treated with Azathioprine initially, in 5 (23%) patients it was necessary to switch to Cyclophosphamide IV due to failure or intolerance. The maintenance treatment was azathioprine in 13 (62%) of the patients, MTX in 4 (19%) and the association of both drugs was indicated in 3 (14%) patients. One case was refractory and received infliximab and subsequently Rituiximab, which resulted in remission.

The best final Visual Acuity (VA) was recorded with the Snellen chart in 40 eyes (20 patients), 27 (67.5%) patients with good vision (20/20 to 20/50), 8 (20%) patients with decreased VA (20/50-20/200), and 5 (12.5%) with legal blindness.

Conclusion: The majority of patients in this series presented a good vision when the inflammatory process was resolved (AV 20/20-20/50), this probably related from early diagnosis and initial aggressive treatment with intravenous corticosteroid pulses and its association with immunosuppressants of entry as mentioned in the literature. The worst vision, (visual acuity <20/200) and of these only 3 patients had unilateral involvement and one patient due to failure or intolerance. The maintenance treatment was azathioprine in 13 (62%) of the patients, MTX in 4 (19%) and the association of both drugs was indicated in 3 (14%) patients. One case was refractory and received infliximab and subsequently Rituiximab, which resulted in remission.

References:

Acknowledgments: Department of Rheumatology

Disclosure of Interests: None declared

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APPLICATION OF THE 2019 ACR/EULAR CLASSIFICATION CRITERIA CONTRIBUTES TO THE DIAGNOSIS OF IGG4-RELATED DISEASE

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Objectives: To evaluate the diagnostic accuracy of the 2019 American College of Rheumatology/European League Against Rheumatism classification criteria for IgG4-related disease.

Methods: This was a retrospective cohort study of patients with IgG4-related disease between 2003 and 2019 who were assessed for the 2019 criteria. The primary endpoint was clinical and epidemiological investigations collected, new classification criteria for
CHARACTERISTICS AND MANAGEMENT OF RHEUMATIC MANIFESTATION UNDER ESTROGEN RECEPTOR-TARGETING CANCER THERAPIES: DATA FROM A PROSPECTIVE REGISTRY

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Background: The knowledge about interdependencies between rheumatic manifestations and malignancies is limited. Further, reliable data on the occurrence of rheumatic symptoms as side effects of specific cancer therapies beyond checkpointinhibitor-induced immune-related adverse events are sparse. In this regard, although arthralgia under estrogen receptor-targeting therapies (aromatase inhibitors and the estrogen receptor modulator tamoxifen) have been frequently reported in oncological clinical trials and case reports, prospective data including an assessment of rheumatic manifestations by rheumatologists are lacking.

Objective: To contribute to a better understanding of interdependencies between rheumatic manifestations and cancer/estrogen blockade and potentially improve the management of both entities, pilot data were analysed.

Methods: Data on characteristics and treatment of rheumatic manifestations and cancer as well as their timely association were systematically, prospectively collected and analysed in the MalheuR (‘malignancy and rheumatic disease’) registry, a long-term, observational study designed to study patients suffering from concomitant rheumatic disease and malignancy and/or premalignant lesions.

Results: We identified 11 patients with rheumatic manifestations under estrogen receptor-targeting therapies (3 anastrozol, 4 letrozol, 8 tamoxifen) as part of breast cancer treatment. In addition to breast cancer one patient had a lymphoma 3 years after and another patient had a non-small cell lung cancer 2 years before breast cancer diagnosis. The patients had different cancer stages (5 IA, 3 IIA, 1 IIB, and 1 IVA). Their mean age at cancer diagnosis was 60.4 ± 11.6 years and all patients are females. The time interval between diagnosis of cancer and onset of systemic/ rheumatic symptoms was 49.5 ± 34.0 months. Of interest, the time interval between onset of rheumatic symptoms and first assessment by a rheumatologist was 16.9 ± 22.3 months. The following systemic and rheumatic symptoms were reported: arthralgia in 10, arthritis in 8 (small joints in 5, large joints in 3 affected), morning stiffness (>30 min) in 7, IIB in 1, myalgia in 7, sicca symptoms in 2, fever in 1 (new-onset CRF with hyperoxygous M694U mutation), class IV (glomerulonephritis), class V (periarteritis nodosa in 1 with new-onset SLE patient) and disease burden at baseline was rather high with a mean VAS pain of 65 (≤12/90). Laboratory analyses revealed an increased CRP in 6/11 (55%) with a mean of 10.3 ± 8.2 mg/L (≤5). Autoantibody positivity was observed for ANAs in 5/10 (50%), titers ranging from 1:80 to 1:160, anti-dsDNA in 1, rheumatoid factor in 4 patients, anti-CCP in 3/11 (27%), with chest involved.

The prospective design of the MalheuR registry enables future validation of our data.